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MODERN CONCEPTS OF CARDIOVASCULAR DISEASE

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Roentgenologic Considerations in the Diagnosis of Congenital Heart Disease (I)*

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With the accumulation of experience in the past decade in the diagnosis and surgical treatment of congenital heart disease, the role of roentgenology has been brought more clearly into focus. While special techniques, such as angiocardiography and aortography, may be essential to precise diagnosis, particularly in complex malformations, conventional radiography remains an important, basic diagnostic tool in the evaluation of patients with congenital anomalies of the heart and great vessels. With improvement in clinical diagnosis and as a result of lessons learned from the correlation of roentgenographic findings with cardiac catheterization, angiocardiographic, surgical and necropsy findings, the interpretation of conventional roentgenograms has become increasingly important and rewarding, especially in view of the trend to treat surgically some of the more simple and uncomplicated malformations without special laboratory study.

While it is important to recognize the value of roentgenology as an aid in the diagnosis of congenital heart disease, it is equally important to appreciate the limitations of this method of diagnosis and to view the place of roentgenography in proper perspective. My assigned task is to discuss some of the capabilities and limitations of the radiologist in the diagnosis of important cardiac malformations and to suggest a useful approach to the analysis of the roentgenologic findings in this group of diseases.

* Part II of this article will appear in the June, 1961, issue.

GENERAL CONSIDERATIONS

It should be unnecessary to say that the contribution of the radiologist is dependent upon his sound knowledge of the basic pathologic, clinical, and physiologic aspects of congenital heart disease and his ability to integrate this knowledge with the roentgenologic findings in a given case. One cannot sufficiently emphasize that the roentgenologic examination is only one link in the chain of methods available for investigation of the cardiovascular system and that only by use of all modes of study is it always possible to assess accurately the anatomic and functional state of the heart. The roentgenologist, using the more routine roentgenologic methods, usually plays only a small, albeit important, contributory role in the diagnosis of congenital heart disease. Although some have estimated that he contributes about 10 per cent to the final diagnosis, it would be more accurate to say that the contribution of the roentgenologic examination varies from case to case. In some conditions, such as coarctation of the aorta or certain types of anomalous pulmonary venous connection, the radiologist can often make a correct diagnosis on the basis of the posteroanterior roentgenogram of the thorax, and his contribution in such instances is large. On the other hand, his

contribution to the diagnosis of more complex anomalies is much smaller, unless special methods of investigation, such as angiocardiography, are used. Thus, there are few times when the roentgenologic findings are characteristic of a specific anomaly; however, on many occasions the radiologist can narrow the diagnosis down to a few specific possibilities, and, by correlating the roentgenologic features with the clinical findings, can suggest which of these several possibilities is most likely.

For example, when the peripheral pulmonary arteries show engorgement of a certain type, the presence of a left-to-right shunt is almost certain. Several diagnostic possibilities, therefore, exist. If the main pulmonary artery is also enlarged and in its normal position, and it is known that the patient is not cyanotic, the possibilities become fewer. If, in addition, the aortic knob is large and at fluoroscopic examination the pulsations of the pulmonary arteries are not greatly accentuated, the chances favor the presence of a patent ductus arteriosus. If, however, the aortic knob is small and the hilar pulsations are markedly increased, the chances favor an intracardiac shunt rather than a patent ductus.

As illustrated by this example, if the roentgenologic examination is to achieve its maximal usefulness, it is important for the radiologist to note on the roentgenogram and at fluoroscopic examination certain pertinent features, which may serve as useful building blocks in the completion of the final diagnostic structure. These pertinent features are the general cardiac configuration; the heart size (as estimated by the cardiothoracic ratio); the size of the individual heart chambers, when it is possible to determine this; the appearance of the main pulmonary artery and its central and peripheral branches; the size, position, and configuration of the aortic arch; and the presence or absence of rib notching. While it is important to note systematically all of these features in each case of suspected congenital heart disease, probably the most important single feature is the appearance of the pulmonary vessels. Not only do changes in the pulmonary circulation reflect the functional pathology of the heart, but the interrelationship is a reciprocal one in that changes in the pulmonary vessels may play a significant role in determining the behavior of the heart. Much helpful information with regard to pulmonary flow and pulmonary vascular resistance may be reflected in the roentgenographic appearance of the pulmonary vessels. This information is of considerable practical importance, both from a diagnostic standpoint and from the standpoint of evaluation of the patient as a candidate for operation.

It is unnecessary to point out to those experienced in this field, but perhaps important to emphasize again in this paper, that there are many traps laid for the unwary in the roentgenologic assessment of congenital heart disease. The difficulties in diagnosis become evident when one remembers that more than one anomaly, and frequently several, may be present in the same patient. Thus, from the roentgenographic findings alone, even when a specific anomaly is evident, it is never possible to be sure that other malformations do not coexist. The need for special laboratory and angiocardiographic study must then depend upon correlation of all available data: clinical, electrocardiographic, and roentgenographic. In addition, it should be emphasized that these various roentgenologic features referred to above are more difficult to evaluate in infants.

CLASSIFICATION

There are several ways of classifying congenital heart diseases. The pathologist classifies them on the basis of embryologic and anatomic features; the cardiologist usually classifies them into cyanotic and acyanotic groups; and the surgeon usually does not classify them at all, because to him each anomaly is a separate and different therapeutic problem. While there is merit in all these methods of classification, or in no classification at all, no one of them is entirely satisfactory to the radiologist.

Knowledge of the presence or absence of cyanosis is, of course, extremely important, for the presence of cyanosis in congenital anomalies, except in terminal heart failure, is usually evidence of a right-to-left shunt or abnormal origin of the great arteries. Although the classification of congenital malformations into cyanotic and acyanotic groups is logical, one must remember that patients with anomalies characterized by a left-to-right shunt and classified in the acyanotic group may, under certain conditions, develop cyanosis. Furthermore, cyanosis is not a roentgenologic finding, and the radiologist, when interpreting roentgenograms, may not have information regarding its presence or absence.

Therefore, a classification has been devised which, from the findings on the conventional

TABLE 1

CLASSIFICATION OF IMPORTANT CONGENITAL MALFORMATIONS

Increased pulmonary vasculature (increased flow):
Ventricular septal defect
Atrial septal defect
Persistent common atrioventricular canal
Patent ductus arteriosus
Aortopulmonary window
Anomalous pulmonary venous connection
Transposition of great arteries
Corrected transposition of great arteries
Truncus arteriosus
Decreased pulmonary vasculature (decreased flow):
Tetralogy of Fallot
Pulmonary stenosis with intact ventricular septum
Pulmonary atresia
Tricuspid atresia
Ebstein's malformation
Agenesis of a main branch of pulmonary artery
Normal pulmonary vasculature (normal flow):
Coarctation of aorta
Congenital aortic stenosis
Pulmonary venous hypertension (obstruction to pulmonary venous flow):
Left heart failure from any cause
Congenital mitral stenosis
Congenital stenosis of pulmonary veins

roentgenogram of the thorax, permits the radiologist to place the patient in one of four broad, diagnostic groups (table 1). This classification is based on changes in the lesser circulation. Thus, patients with congenital heart disease are divided into three groups, depending upon whether there is increased, decreased, or normal pulmonary arterial vasculature. Increased pulmonary vasculature, and therefore increased flow, is seen in patients with a left-to-right shunt. Decreased pulmonary vasculature, or decreased flow, is seen in patients with obstruction to pulmonary arterial flow and a right-to-left shunt. Normal pulmonary vasculature is, in general, seen in a miscellaneous group of malformations in which there is no direct communication between the systemic and lesser circulations and, therefore, no shunt or admixture of arterial and venous blood. A fourth group to be considered consists of congenital lesions producing pulmonary venous hypertension resulting from obstruction of pulmonary venous flow.

Although this suggested classification shares some of the weaknesses of that based on the presence or absence of cyanosis, it would seem a more logical approach from the standpoint of roentgenographic interpretation.

CONDITIONS CHARACTERIZED BY INCREASED PULMONARY VASCULATURE

In the group of conditions characterized by increased pulmonary vasculature, a number of cardiac anomalies must be considered. In each of these anomalies, there are functionally a left-to-right shunt, either intracardiac or extracardiac, and increased pulmonary artery flow.

The most commonly encountered anomalies in this group are ventricular septal defect, atrial septal defect, patent ductus arteriosus, persistent common atrioventricular canal, and anomalous pulmonary venous connection. These five conditions with left-to-right shunt comprise more than 50 per cent of all cases of congenital heart disease. Also included in this group with increased pulmonary flow are four other important, but less common, malformations: aortopulmonary window (partial truncus), transposition of the great arteries, corrected transposition of the great arteries, and persistent truncus arteriosus.

The chief roentgenographic characteristic of all of the malformations in this group with left-to-right shunt, with the exception of transposition, corrected transposition, and truncus, is increase in the size of the main pulmonary artery and its central and peripheral branches, as evidence of increased pulmonary flow. In transposition, corrected transposition, and truncus,

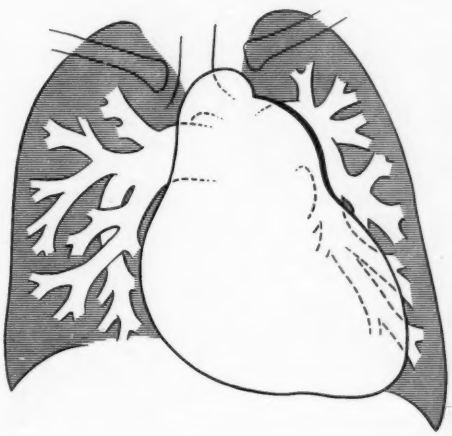

Figure 1. *Ventricular septal defect with large left-to-right shunt. Heart markedly enlarged. Main pulmonary artery and its hilar and peripheral branches are greatly enlarged. Aortic knob is small.*

the findings are somewhat different, although there is usually increased pulmonary vasculature in these malformations also. Estimation of ventricular enlargement and left atrial size is of little practical help in differentiating between the various defects associated with left-to-right shunt.

Ventricular Septal Defect, Atrial Septal Defect, Common Atrioventricular Canal, Patent Ductus, Aortopulmonary Window

There are no distinct roentgenographic differences between ventricular septal defect, atrial septal defect, and persistent common atrioventricular canal (fig. 1).* In addition to increased pulmonary vasculature, these three conditions are commonly associated with a hypoplastic aortic knob and vigorous pulsation of the pulmonary arteries as seen fluoroscopically.

Patent ductus arteriosus, however, often produces a somewhat different picture in that the aortic knob is not hypoplastic and frequently is bulbous in appearance (fig. 2), and the central pulmonary arteries are often less enlarged and at fluoroscopic examination are seen to pulsate less than in patients with large left-to-right intracardiac shunts. This lesser pulsation is probably due to the fact that the flow of blood from the systemic to the pulmonary circulation occurs continuously in uncomplicated patent ductus, but intermittently in the intracardiac shunts. In possibly 50 per cent of cases, the radiologist is able to distinguish between patent ductus and other lesions with left-to-right shunt on the basis of these differentiating features. However, this distinction can rarely be made in infants and younger children, since the aortic knob in these age groups is seldom clearly discernible.

In aortopulmonary window (partial truncus), the aortic knob is usually not bulbous, as it is in patent ductus, and, although the pulmonary artery and its hilar branches do not pulsate as vigorously as in patients with septal defects or common atrioventricular canal, differentiation from these latter conditions is not possible.

* Line drawings for this article were made from actual roentgenograms by Mr. Vincent Destro. Heavy lines in each instance are for the purpose of emphasizing certain important features.

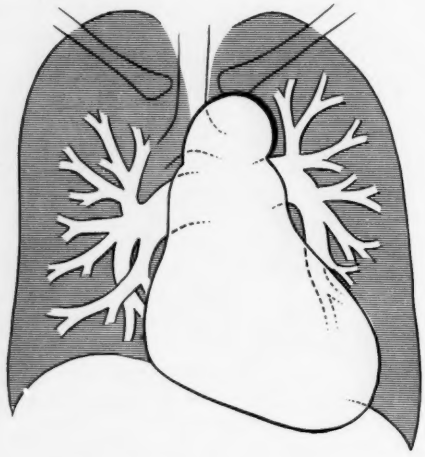

Figure 2. *Patent ductus arteriosus. Moderate enlargement of heart and of main pulmonary artery and its hilar and peripheral branches. Note large bulbous aortic knob.*

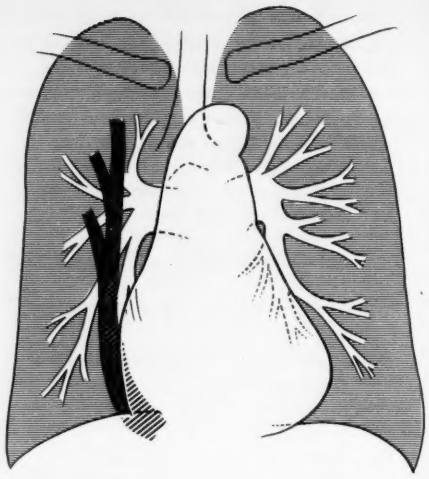

Figure 3. *Anomalous venous connection of right lung to inferior vena cava. Note large abnormal pulmonary vein coursing downward along right heart border. Pulmonary arterial vasculature in this case is normal or perhaps only slightly increased. Cardiac silhouette is normal.*

Anomalous Pulmonary Venous Connection

In anomalous pulmonary venous connection, one or more of the pulmonary veins may drain anomalously into the right atrium, superior vena cava, inferior vena cava, hepatic vein, portal vein, coronary sinus, left innominate vein, persistent left superior vena cava, ductus venosus, or azygos vein. This condition is often present without evidence of the anomalous vein or veins on the roentgenogram. In most cases, the only roentgenographic abnormality is prom-

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MORNING	SESSIONS ON CLINICAL CARDIOLOGY	Address by President Conner Memorial Lecture Symposium: Contribution of Phonocardiography to Auscultation	Panel: Ventricular Arrhythmias Closed Chest Cardiac Resuscitation	Jointly with Basic Science Symposium: Role of Hormones in Heart Failure Venous Factors in Control of the Circulation
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		High Blood Pressure Research	Community Service and Education Compensable Heart Disease, Strain and Trauma	Cardiovascular Films
AFTERNOON	SESSIONS ON CLINICAL CARDIOLOGY	Cardiovascular Radiology Symposium: Coronary Arteriography Biplane Angiography	Jointly with Circulation Brown Memorial Lecture Symposium: Renal Failure	Panel: Ventricular Hypertrophy and Bundle Branch Block Panel: Newer Electrocardiographic Lead Systems ECG Clues Suggesting Myocardial Infarction
	SPECIAL AND COUNCIL SESSIONS	Arteriosclerosis	Basic Science	Rheumatic Fever and Congenital Heart Disease
		Circulation	Cardiovascular Surgery Jointly with Rheumatic Fever and Congenital Heart Disease	Community Service and Education Session for Public
		Community Service and Education Session for Nurses	Community Service and Education Compensable Heart Disease, Strain and Trauma	Cardiovascular Films
	EVENING		Cardiac Conferences	Annual Dinner American Heart Association

Sunday, October 22 — 2:00 P.M.	Program for Assembly Delegates
Monday, October 23 — all day	Assembly Panel Meetings
12:15 P.M.	Assembly Luncheon
Tuesday, October 24 — 9:00 A.M. — 12 Noon	Annual Meeting of Assembly, elections — panel reports.

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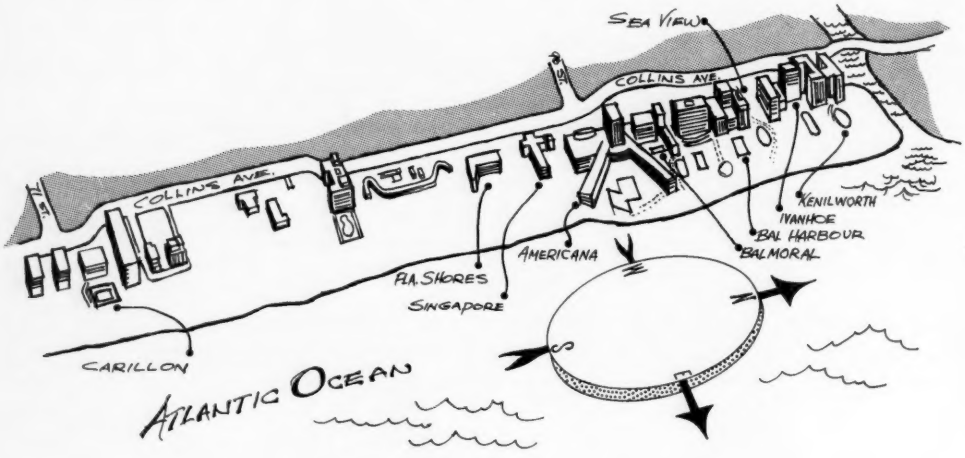

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The Heart Association of Greater Miami and the Florida Heart Association will be your hosts during the American Heart Association Annual Meeting. The Eastward Room is designated as the "Heart Hospitality Room" at the Americana Hotel and will be open from Friday, October 20 through Monday, October 23.

Hostesses will be there to greet you, to serve coffee and fresh orange juice and to help in every way to make your stay in Miami Beach a pleasant one. Please use the "Heart Hospitality Room" to meet your friends.

The following special events have been planned. More detailed information will be available later.

FRIDAY, OCTOBER 20

10:00 A.M.	Opening of the Heart Hospitality Room. Special Open House from 3:00 to 5:00 P.M. for meeting and greeting friends.

SATURDAY, OCTOBER 21

12:30 P.M.-3:00 P.M.	Luncheon and Fashion Show at the fabulous Fontainebleau Hotel, Miami Beach.

SUNDAY, OCTOBER 22

12:30 P.M.-3:30 P.M.	Scenic boat trip around beautiful waterfront residences—including buffet luncheon on board boat.

MONDAY, OCTOBER 23

MORNING	Tour of tropical Parrot Jungle, paradise of beautiful birds in a spectacular show and visit to Fairchild Gardens—a wonderland of lush tropical foliage and interesting trees, seen only in the tropics, or visit to Villa Vizcaya, magnificent Italian palace in a tropical setting.
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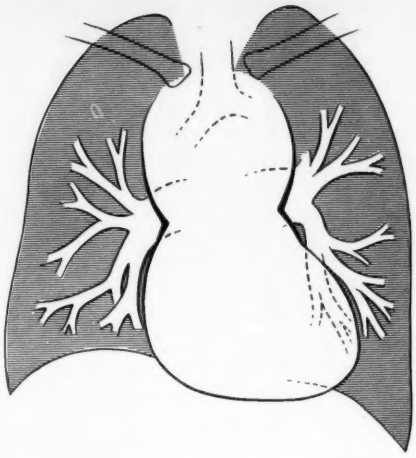

Figure 4. *Total anomalous pulmonary venous connection to a persistent left superior vena cava. Note figure-of-8 silhouette. Heart is slightly enlarged. Central and peripheral pulmonary artery branches are prominent. Aortic knob is obscured.*

inence of the central and peripheral pulmonary arteries, due to the pulmonary recirculation. In these cases, it is impossible to differentiate anomalous pulmonary venous connection from the other anomalies characterized by a left-to-right shunt.

There are, however, two types of anomalous pulmonary venous connection that present characteristic roentgenographic findings. In the first, all or part of the right lung drains anomalously into the inferior vena cava, hepatic vein, or portal vein below the right hemidiaphragm. In such cases, the anomalous vein may be seen coursing downward along the right cardiac border (fig. 3). Tomography is helpful in more clearly defining the anomalous vein. This type of anomalous venous connection is often associated with hypoplasia of the involved lung and with an anomalous arterial supply to this lung from the descending aorta, resulting in an unusual shift of the heart into the right hemithorax.

In the other type, there is total anomalous venous connection with the veins from both lungs draining into a persistent left superior vena cava. The striking and characteristic feature of this anomaly is the so-called figure-of-8 conformation, with the heart forming the lower half of the "8" and the large left and right superior venae cavae forming the upper half (fig. 4).

Transposition of the Great Arteries

Three other important malformations that are characterized by increased pulmonary vasculature owing to increased pulmonary flow but that have roentgenographic features often distinctly different from the conditions just discussed are transposition of the great arteries, corrected transposition of the great arteries, and persistent truncus arteriosus.

In transposition of the great arteries, increased pulmonary vasculature is consistently present. However, the pulmonary artery segment of the left heart border is very often flat or concave, rather than prominent and convex (fig. 5). This particular finding of a concave pulmonary artery segment with increased peripheral vasculature is the most consistent and characteristic roentgenographic feature of transposition. In those cases of transposition in which the aorta lies directly anterior to the pulmonary artery, the supracardiac vascular pedicle may appear abnormally narrow in the posteroanterior view. This latter finding, however, is lacking in the majority of cases of transposition.

Corrected Transposition of the Great Arteries

In that anomaly known as corrected transposition of the great arteries, the pulmonary vasculature is increased owing to the fact that a ventricular septal defect or absence of the

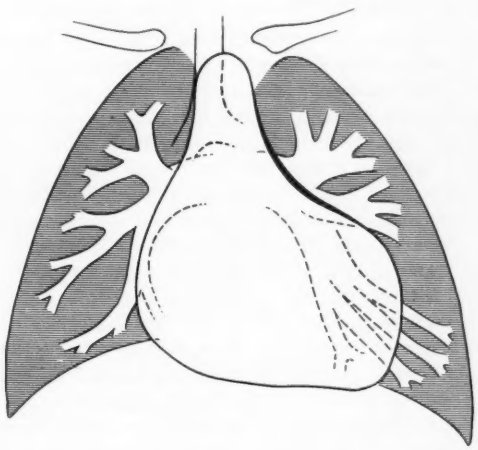

Figure 5. *Complete transposition of the great arteries with ventricular septal defect. Note slightly concave pulmonary artery segment but prominent peripheral pulmonary vasculature. Supracardiac vascular pedicle is narrow. Marked cardiac enlargement.*

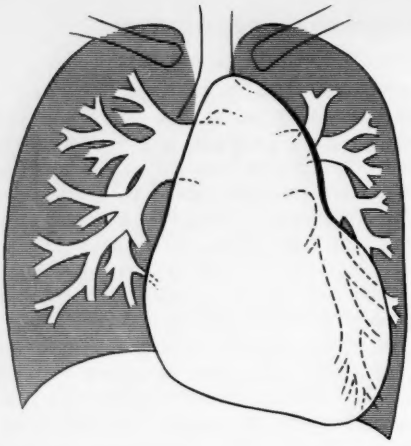

Figure 6. *Corrected transposition with ventricular septal defect. Long prominent convex upper left border produced by ascending aorta. Since septal defect is present, central and peripheral pulmonary artery branches are enlarged.*

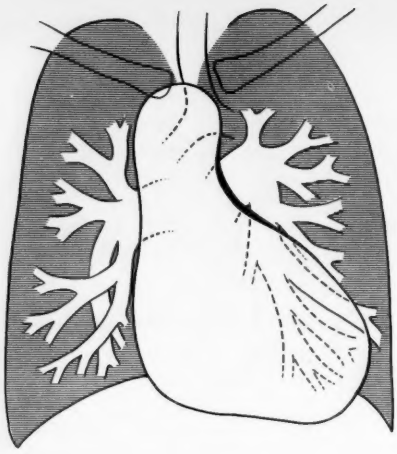

Figure 7. *Truncus arteriosus. Pulmonary artery segment flat or slightly concave, but peripheral vasculature quite prominent. Right and left main branches of pulmonary artery are higher than normal and do not appear to have a normal connection to the outflow region of the right ventricle. Aortic shadow is broad and aortic arch is on the right. Heart is moderately enlarged.*

ventricular septum is commonly associated with this condition. The diagnosis of corrected transposition of the great arteries can be made with considerable accuracy in many cases because of the characteristic position of the ascending aorta, which arises on the left side and overlies the pulmonary artery segment of the left heart border (fig. 6).

Truncus Arteriosus

In persistent truncus arteriosus, the pulmonary artery does not arise in normal fashion from the right ventricle, but instead arises from a large common trunk overriding a high ventricular septal defect and guarded by a single valve. This anomaly can be suspected on the roentgenogram whenever the aortic shadow appears wider than normal and the pulmonary arteries are prominent but do not appear to have a normal connection with the outflow tract of the right ventricle (fig. 7). In this condition, the pulmonary vasculature is increased but, as in transposition of the great arteries, the pulmonary artery segment is often flat or concave, a finding of great importance in differentiating these from the other anomalies with increased pulmonary vasculature. An additional feature of persistent truncus arteriosus is that the aortic arch is on the right in approximately 25 per cent of cases.

Angiocardiography is usually necessary to make, with certainty, the diagnosis of transposition or corrected transposition of the great arteries and of persistent truncus arteriosus.

Roentgenography as an Aid in Surgical Evaluation

As mentioned earlier, the radiographic findings in patients with congenital heart disease are important not only from a diagnostic standpoint but also from the standpoint of evaluation of operability. One reason the early diagnosis of ventricular septal defects is important is that in patients with this condition and a large left-to-right shunt, obliterative changes may soon occur in the smaller pulmonary arteries and increase peripheral pulmonary vascular resistance to the point where little or no left-to-right shunt occurs. As these changes in the small peripheral arteries progress, the shunt becomes bidirectional and may become predominantly right to left. Patients in whom this increased-resistance type of pulmonary hypertension has developed may not be benefited by surgical closure of the defect, and operation may be disastrous, because the changes in the small pulmonary arteries, when sufficiently advanced, may be irreversible. When the defect is closed in these patients, pressure in the pulmonary artery remains elevated, due to the increased resistance present in the pulmonary vascular bed, and the augmented work load of maintaining the pulmonary circu-

lation placed on the right ventricle may cause it to fail. These changes in the small pulmonary arteries occur most commonly in ventricular septal defect, since in this condition the pulmonary arteries are functionally in direct communication with the left ventricle, a high-pressure chamber. Such changes may also occur, however, in patent ductus arteriosus or, less frequently, in any of the other conditions in which there is increased pulmonary flow.

The presence of this increased-resistance type of pulmonary hypertension may often be recognized on the posteroanterior roentgenogram of the thorax. In defects with a large left-to-right shunt, and therefore a large pulmonary-to-systemic flow ratio, the central pulmonary arteries are prominent and the peripheral pulmonary arteries are enlarged all the way out to the periphery of the lung. In patients who have developed increased peripheral pulmonary vascular resistance owing to intimal changes in the smaller pulmonary arteries and, thus, reduction in pulmonary-to-systemic flow ratio, the peripheral vessels are seen to be narrowed or normal in size while the central pulmonary arteries are enlarged. Thus, the roentgenographic recognition of this situation is based on the presence of enlargement of the central pulmonary arteries and disproportionate narrowing or tapering of the peripheral pulmonary arteries (fig. 8).

It should be pointed out, however, that while one is able to recognize quite easily, on the one hand, those patients with a large pulmonary-to-systemic flow ratio, and, on the other hand, those with marked pulmonary vascular disease and a reduced pulmonary-to-systemic flow ratio, a large borderline group exists in which evaluation of the pulmonary-to-systemic flow ratio on the basis of the roentgenographic appearance of the pulmonary vessels may be quite inaccurate.

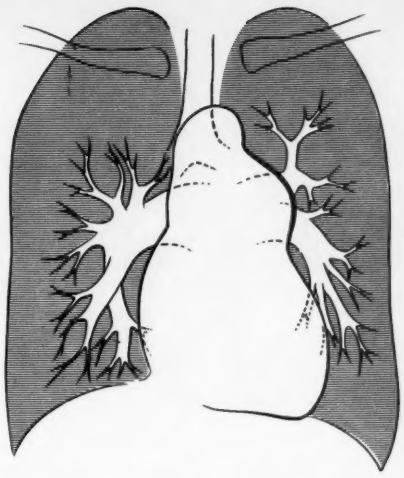

Figure 8. *Ventricular septal defect following development of pulmonary vascular disease. There are now a predominant right-to-left shunt and cyanosis. Main pulmonary artery and its hilar branches are markedly enlarged, but peripheral pulmonary arteries taper quite rapidly and pulmonary vasculature in outer third of lung fields is decreased from normal. Heart is now less enlarged, since there is no longer great left ventricular overload.*

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